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Stratifying connective tissue diseases

SP0173 STRATIFYING PRIMARY SJÖGREN'S SYNDROME PATIENTS

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The clinical manifestations of primary Sjögren's syndrome (PSS) vary considerably between individual patients. The pathogenesis of PSS is not fully understood and although many aberrant biological pathways have been identified, the relationships between these pathways and clinical manifestations remain unclear. Furthermore, there is substantial heterogeneity in long-term outcomes and in health economic cost among patients with PSS. Stratification of PSS into more homogeneous subsets will facilitate better understanding of the pathogenesis of the disease, development of more cost-effective management strategies and accelerate therapeutic development programmes of targeted therapies in PSS. The establishment of several large cohorts of clinically well characterised PSS cohorts with associated biobanked materials has provided a rich resource for stratified medicine research in PSS. The growing number of datasets from clinical trials in PSS provide an excellent opportunity to evaluate the potential impact of stratified approach to therapy development in PSS.

In this presentation, different approaches to identify clinically and biologically meaningful PSS subsets will be considered. In addition, the key challenges and potential future directions in Stratified Medicine in PSS will be discussed.

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SP0174 THE VALUE OF TYPE I INTERFERON SIGNATURE TO STRATIFY PATIENTS

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Since the first reports 2003 of an increased expression of type I interferon (IFN) regulated genes in cells from patients with SLE, a number of studies have demonstrated that many systemic inflammatory autoimmune diseases display this over-expression of type I IFN regulated genes, now known as an IFN signature. This is

true for a majority of patients with SLE, but also large proportion of patients with primary Sjögren's syndrome, myositis, systemic sclerosis and rheumatoid arthritis (RA). The so called type I interferonopathies is a group of rare Mendelian diseases with a very strong IFN signature. Other autoimmune diseases, such as type I diabetes, also display an IFN signature. Type I IFN seems to be most important for inducing the IFN signature, but it's clear that type II and type III IFNs can contribute to the IFN signature. Besides the expression of type I IFN regulated genes, recent studies have shown that patients with SLE and other autoimmune disease, have epigenetic changes in IFN regulated gene, which are hypomethylated. Thus, there are strong evidences that type I IFN system activation is a key event in many autoimmune diseases. Can the IFN signature help us to stratify patients in order to predict risk for major organ manifestations, disease outcome or response to treatment? Early studies showed that patients with active disease and more severe disease manifestations usually have a more prominent IFN signature, which still is true. However, recent studies have revealed that the IFN signature needs to be combined with a broader analysis of expressed genes. For instance, a plasmablast signature seems to correlate best with SLE disease activity and a neutrophil signature associates with lupus nephritis. For treatment stratification, it's perhaps not surprising that SLE patients with a high IFN signature have a better treatment response to type I IFN inhibition, compared to IFN low patients. At least when type I IFN receptor inhibition has been used to downregulated the type I IFN response. In RA, the IFN signature at baseline predict a poor response to TNF-alpha treatment as well as nonresponse to B-cell depleting therapy. In conclusion, there are several indications that the type I IFN signature is a valuable biomarker for patient stratification. However, we are just at the brink of the understanding how to apply this signature in clinical practice.

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SP0175 HETEROGENEITY OF MYOSITIS AND THERAPEUTIC CONSEQUENCES

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Idiopathic inflammatory myopathies, or myositis, are a heterogeneous group of chronic inflammatory disorders affecting striated muscle leading to muscle weakness and impaired function. In addition, other organs are variably involved including the skin, lungs, heart, joints, and the gastrointestinal tract contributing to morbidity and mortality and to low health-related quality of life. Treatment decisions are often guided by the most prominently affected organ. The best recognised disease subsets are polymyositis (PM), dermatomyositis (DM), juvenile PM and DM, sporadic inclusion body myositis (sIBM), and immune-mediated necrotizing myopathy (IMNM). Muscle specific autoantibodies and muscle associated autoantibodies are frequently found in different forms of myositis and collectively they can be detected in approximately 80% adults and 60% children with myositis. They can identify homogeneous subsets, and have both diagnostic and prognostic value. The worst functional outcomes in terms of muscle involvement are often seen in patients with sIBM and with anti-SRP antibodies. sIBM patients are notoriously non-responsive to any treatment. Patients with anti-SRP display necrotizing myopathy in the muscle biopsy and treatment effect is usually low. In some patients an improvement or at least stabilisation can be achieved with the use of rituximab. The other patients with IMNM are those with anti-HMGCR antibodies and with statin medication history. Here the response to glucocorticoids (GC) and immunosuppressives (IS) is mostly good, although some patients require treatment with intravenous immunoglobulins (IVIg) to which they respond very well even without concomitant GC. Patients with antisynthetase syndrome are characterised by the presence of interstitial lung disease, which may be in some cases the prevailing manifestation of the disease with only mild or none muscle weakness. Treatment is often successful with calcineurin inhibitors, mycophenolate mofetil (MMF), cyclophosphamide, or rituximab in these patients. Subgroup of patients with anti-NXP2 antibodies is characterised with strong association with calcinosis, both in children and adults, and the treatment represents a difficult problem. The effect was reported with the use of calcium channel blockers, warfarin, bisphosphonates, sodium thiosulfate, aluminium hydroxide, probenecid, colchicine as well as IVIGs, rituximab, abatacept, infliximab, and thalidomide. In any case, patients need to be treated aggressively if calcinosis progresses. Muscle disease in dermatomyositis is often responsive but the treatment of cutaneous manifestations may occasionally prove difficult. Antimalarials are usually prescribed first, sometimes in combination with systemic treatment using IVIGs, tacrolimus, MMF, or other IS. Patients with anti-MDA5 antibodies often develop clinically amyopathic or hypomyopathic DM, but at the same time their ILD can be rapidly progressive. Aggressive treatment with high dose GC and IS needs to be started early to increase the chance for survival. A proportion of patients with myositis manifests high type I interferon signature and the clinical trial suggests that these are the patients who benefit most from the treatment with interferon type I inhibition. In the future careful considerations based on the disease subsets,